



## General

### Guideline Title

ACR Appropriateness Criteria® known or suspected congenital heart disease in the adult.

### Bibliographic Source(s)

Ho VB, Biko DM, White RD, Woodard PK, Abbata S, Dorbala S, Haramati LB, Mammen L, Martin ET III, Rigsby CK, Ryan T, White CS, Expert Panel on Cardiovascular Imaging. ACR Appropriateness Criteria® known or suspected congenital heart disease in the adult. [online publication]. Reston (VA): American College of Radiology (ACR); 2011. 9 p. [80 references]

### Guideline Status

This is the current release of the guideline.

This guideline updates a previous version: Ho VB, Yucel EK, Khan A, Haramati LB, Mammen L, Rozenshtein A, Rybicki FJ, Schoepf UJ, Stanford W, Stein B, Woodard PK, Jaff M, Expert Panel on Cardiovascular Imaging. Suspected congenital heart disease in the adult. [online publication]. Reston (VA): American College of Radiology (ACR); 2007. 8 p.

The appropriateness criteria are reviewed biennially and updated by the panels as needed, depending on introduction of new and highly significant scientific evidence.

## Recommendations

### Major Recommendations

ACR Appropriateness Criteria®

Clinical Condition: Known or Suspected Congenital Heart Disease in the Adult

Radiologic Procedure	Rating	Comments	RRL*
US echocardiography transthoracic resting	9		O
MRI heart function and morphology without and with contrast	8	See statement regarding contrast in text under "Anticipated Exceptions."	O
Catheterization with angiography	4,5,6 May be appropriate;	7,8,9 Usually appropriate. Adjunctive to noninvasive imaging for hemodynamic measurements and when intervention is needed.	*Relative Radiation Level

			<input type="text"/>
X-ray chest	7		<input type="text"/>
MRI heart function and morphology without contrast	7		O
CT heart function and morphology with contrast	7	May be alternative to MRI and TTE/TEE. High spatial resolution to evaluate small and tortuous vessels. For evaluation of airway.	<input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/>
US echocardiography transesophageal	6	May be appropriate as an adjunct to TTE for further evaluation of valves and septal defects.	O
MRA chest (noncoronary) without contrast	6	For great-vessel assessment and to evaluate associated vascular abnormalities.	O
MRA chest (noncoronary) without and with contrast	6	For great-vessel assessment and to evaluate associated vascular abnormalities. See statement regarding contrast in text under "Anticipated Exceptions."	O
CTA coronary arteries with contrast	6	When there is a high index of suspicion for fistula or anomalous coronaries.	<input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/>
CTA coronary arteries with contrast with advanced low dose techniques	6	Important in young to middle-age adults because of reduced radiation dose.	<input type="text"/> <input type="text"/> <input type="text"/>
Tc-99m ventriculography	2		<input type="text"/> <input type="text"/> <input type="text"/>
<u>Rating Scale:</u> 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate			*Relative Radiation Level

Note: Abbreviations used in the table are listed at the end of the "Major Recommendations" field.

#### Summary of Literature Review

Congenital heart disease (CHD) has been estimated to occur in approximately 0.4% to 1% of live births. The number of adult patients in North America with CHD has increased over the past three decades. Although this increase is attributable in part to growth of the adult population, it also results from a number of other causes, notably improvements in surgical treatment that have translated to improved survival, which is greater than 90% at 10 years. A review of data from the National Center for Health Statistics and the Centers of Disease Control and Prevention showed a 39% decline in reported mortality from CHD in the United States between 1979 and 1997. In North America, more than 500,000 adults have had surgically treated CHD, with nearly 50% requiring two or more operations and 23% requiring three or more. The increasing prevalence of adult CHD and its rise in health care utilization are demonstrated by the doubling of adult CHD hospitalizations in the U.S. between 1998 and 2005. In particular, patients with more severe CHD (tetralogy of Fallot, truncus arteriosus, transposition complexes, endocardial cushion defects, and univentricular hearts) are living longer such that the numbers of adults and children alive with severe CHD have become nearly equal over the past decade. The recent Canadian Cardiovascular Society 2009 Consensus conference suggests that there are currently more adults with CHD than children.

Congenital heart lesions may become symptomatic at any time from birth until adulthood. Several common congenital heart defects often survive into adulthood. These include bicuspid aortic valve, congenital forms of mitral valve prolapse, aortic coarctation, atrial septal defect (ASD), pulmonary valve stenosis, patent ductus arteriosus, and tetralogy of Fallot. Uncommon congenital cardiac defects that may present in adulthood

include Ebstein's anomaly, corrected transposition of the great vessels, pulmonary arteriovenous malformation, coronary artery anomalies, and sinus of Valsalva aneurysm. Approximately 10% of patients with common CHD survive undetected until adulthood. The most common congenital heart defect in children, ventricular septal defect (VSD), may escape detection and present in adults either as a small, physiologically insignificant defect or as a large defect with Eisenmenger physiology. Anomalies of the great arteries such as complete transposition and total anomalous pulmonary venous drainage are usually symptomatic, whereas less significant anomalies such as a persistent left superior vena cava and many anomalies of the origin of the great vessels from the arch are often asymptomatic.

Imaging procedures for the diagnosis of known or suspected CHD in the adult include plain chest film radiography, fluoroscopy, echocardiography (transthoracic and transesophageal), nuclear scintigraphy, cardiac-gated computed tomography (CT), magnetic resonance imaging (MRI), and cardiac catheterization and angiography. The physician trying to diagnose these often-complex conditions needs complete and reliable information that includes details about intercardiac anatomy, vascular anatomy, hemodynamics, and function. Adults with CHD also have acquired comorbid factors related to aging, such as hypertension, atherosclerosis, coronary artery occlusive disease, pulmonary disease and renal disease, which may complicate their medical and/or surgical management. When comorbid factors are present and in some congenital conditions (e.g. anomalous coronary artery, univentricular heart), the capability of the myocardium to meet metabolic and physiologic needs may be uncertain. In these instances, reference should be made to the ACR Appropriateness Criteria® related to the evaluation of myocardial ischemia (see the National Guideline Clearinghouse [NGC] summary of the [Chest Pain, Suggestive of Acute Coronary Syndrome](#) and [Chronic Chest Pain – High Probability of Coronary Artery Disease](#) [ ]).

### Chest Radiography

The initial work-up of adults with known or suspected CHD usually includes a posteroanterior and lateral chest radiograph. Occasionally the radiograph will be the first study to alert the radiologist and the clinician to the possibility of a congenital cardiac defect or great-vessel anomaly. This simple and inexpensive examination remains a first-line test for patients with suspected CHD.

The chest radiograph quickly illustrates gross cardiac and mediastinal contours, pulmonary vascularity, pathologic calcification, and the presence of certain indwelling metallic devices. It also provides an assessment of cardiac size, cardiac configuration, and position of the aortic arch. The situs of the abdomen and thorax can usually be determined. Thoracic cage anomalies associated with CHD and postoperative changes may also be detected. The chest radiograph continues to be a useful tool for the initial assessment of the patient with surgically treated CHD.

### Transthoracic Echocardiography

Transthoracic echocardiography (TTE) remains a first-line imaging examination in adults with known or suspected CHD. This test has long been established as a clinically useful diagnostic modality for CHD in children, often eliminating the need for cardiac catheterization in uncomplicated lesions. Although adults present certain technical problems related to the need for lower frequency transducers, limited acoustical windows, and postoperative changes, this examination provides a unique, 2-dimensional (2D), real-time evaluation of the anatomic and hemodynamic relationships of intracardiac lesions.

TTE is widely available, reproducible, safe, and painless. As such it remains a valuable tool in the investigation of CHD.

Echocardiography using color flow Doppler is essential for evaluating blood flow as seen across an atrial defect or a VSD or across a valve. Assessment of the valves (sclerosis, fusion, estimation of valve gradients) and determination of right ventricular systolic pressure can usually be achieved.

TTE, however, has difficulty in consistently providing high-quality clinically useful information in some adult patients with intracardiac defects. Imaging of the great vessels with TTE is difficult even in children and is even more problematic in adults who have poorer acoustical windows. In these situations, transesophageal echocardiography (TEE) and MRI have roles to play. Echocardiography also suffers from intraobserver variability in terms of examination reproducibility.

Current 2D TTE is limited by a field of view of 90 degrees and the need for the examiner to assimilate tomographic slices into a 3- or 4-dimensional diagnosis. The recent development of a rotational acquisition format with dynamic volume rendering has allowed presentation of TTE in a 3-dimensional (3D) display. In one study, 3D echocardiography was found to be particularly useful, when compared to 2D echocardiography, for evaluating mitral valve, aortoseptal continuity, and the intra-arterial septum. While 3D echocardiography can provide more information than traditional 2D techniques, it has been reported to be nondiagnostic in up to 27% to 48% of patients secondary to inadequate quality from morbid obesity, narrow intercostal spaces, and severe pulmonary emphysema.

### Transesophageal Echocardiography

TEE has clear advantages over TTE in adolescents and adults with CHD. TEE can provide a new or altered diagnosis (14%) or new information (56%) in adults with CHD. New information obtained with TEE as compared to TTE includes identification of the atrial appendages and atrial

septum, delineation of systemic and pulmonary venous connections, improved morphologic assessment of the atrioventricular junction and valves, improved definition of subaortic obstruction, improved definition of the ascending aorta and coronary arteries, and better evaluation of atrial baffle function and Fontan anatomy. Limitations of TEE include limited planes of view, poor visualization of specific regions (e.g., apical-anterior septum and right ventricular outflow tract), and blind areas created by masking of flow by implanted prosthetic material. Areas that may be difficult to visualize on TEE are the right ventricular outflow tract, the pulmonary valve, the distal right pulmonary artery, and the proximal left pulmonary artery. With the addition of the vertical axis in the newer probes, these problem areas, as well as the pulmonary veins, are better seen.

The standard TEE is an invasive examination that requires administration of a local anesthetic to the pharynx and intravenous midazolam in small doses. In large studies, it has been shown that the examination may be unsuccessful in up to 5% of patients due to their inability to tolerate the probe after intubation. Another 4%-5% of patients have the examination while under general anesthesia as part of invasive or surgical procedures. Although the risk of bacterial endocarditis from TEE is small, and prophylactic antibiotics are not routinely administered, endocarditis has been attributed to the procedure.

TEE is clearly operator dependent. In an area as complex as CHD, the examiner must be trained to interpret the findings in real time so that important information is not missed.

### Radionuclide Imaging

Although quantitation of cardiac shunts is feasible using technetium Tc 99m first-pass techniques, it is seldom used today. There are, however, a few selected uses for radionuclide imaging in evaluating adults with CHD. Left ventricular dysfunction is known to complicate certain long-standing congenital heart defects associated with right and left heart volume overload. Left ventricular radionuclide scintigraphy with ejection fraction calculation can be a useful noninvasive technique for evaluating these patients. Gated radionuclide scintigraphy can improve risk assessment in patients with single or systemic right ventricles by assessing heart failure and ventricular dysfunction. In some adult patients with abnormal pulmonary blood flow patterns related to conditions such as pulmonary artery agenesis, perfusion lung scanning may assist in the diagnosis. Congenital anomalies of the coronary artery origins, notably anomalous origin of the coronary artery from the pulmonary artery and interarterial anomalous coronary artery, may result in myocardial ischemia and/or silent infarction, which can be identified using stress/rest radionuclide single photon emission computed tomography (SPECT) imaging. Stress/rest radionuclide SPECT imaging can also be used to evaluate myocardial perfusion and function of the systemic right ventricle in patients following repair of transposition of the great vessels, in which perfusion defects can commonly (54%) be seen on long-term follow-up.

### Computed Tomography

Cardiac-gated CT and CT angiography (CTA) can contribute valuable information about congenital abnormalities of the coronary arteries and thoracic aorta, including the identification of vascular rings and postoperative complications such as pseudoaneurysm. CT is widely available and has a short acquisition time, reducing the need for sedation. The exposure to radiation, however, has limited the use of CT in the pediatric population with CHD. It requires the use of contrast media with their potential for adverse effects, but such effects occur less often with the newer low-osmolality agents.

Current CT scanners can evaluate the entire heart and great vessel region in a 3D matrix of CT information. Cardiac gating is not always necessary but it is needed for evaluating small intracardiac connections and coronary artery anomalies, and to detect cardiac isomerism. Essentially all types of congenital cardiac malformations have been accurately described using cardiac-gated CT. Cardiac-gated CT has been used to calculate cardiac output, shunt flow, pulmonary-to-systemic flow ratios, ventricular volumes, ejection fraction, regurgitant volumes, and myocardial mass. CT is limited in evaluation of the interatrial septum and membranous portion of the ventricular septum and is not currently used to assess flow patterns and turbulence.

CT is advantageous in its ability to evaluate extracardiac anatomy such as the airway, sometimes eliminating the need for more invasive conventional tracheobronchoscopy. Compression of the airway may be present in CHD due to abnormal enlargement or an abnormal path of vasculature. Thin-section virtual tracheobronchoscopic images using surface rendering technique enables 3D and comprehensive evaluation of the airway. Inspiratory and expiratory CT imaging is useful in evaluating tracheomalacia in patients with vascular anomalies. Additionally, due to its high spatial resolution, CT is useful in assessing smaller and tortuous vessels as commonly seen in disorders such as total and partial anomalous pulmonary venous return and tetralogy of Fallot with pulmonary atresia. CT can also be used as an alternative to MRI in patients with contraindications to MRI and in patients with metallic implants that may limit the use of MRI.

Recent advances in cardiac CT imaging technology allow for further reduction of the radiation dose from traditional coronary CTA (CCTA) techniques. Available new dose-reducing techniques include prospective triggering, adaptive statistical iterative reconstruction, and high-pitch spiral acquisition. These techniques provide the anatomic detail needed to evaluate many congenital heart diseases.

### Magnetic Resonance Imaging

MRI is invaluable for evaluating CHD. Without the concerns related to exposure to ionizing radiation or the use of iodinated contrast agents, it can provide morphologic and functional information essential for detecting and managing CHD. Traditional "black-blood" techniques (e.g., spin-echo MRI and double inversion recovery fast spin echo) are useful for delineating basic cardiac and pericardiac anatomy. "Bright-blood" techniques, notably using newer cine steady-state free-precession pulse sequences, can demonstrate flow abnormalities (e.g., a flow jet) related to lesions such as an interventricular or interatrial septal defect, valvular insufficiency, valvular stenosis, outflow obstruction, or aortic coarctation. Parallel imaging and newer k-space schemes can shorten the acquisition times in most instances such that cine bright-blood imaging can be performed during a short breath hold. Bright-blood techniques also enable volumetric coverage of cardiac chambers for determining cardiac metrics such as ventricular volumes, ejection fractions, and myocardial mass. Longer acquisitions as may be required for coronary magnetic resonance angiography (MRA) are typically performed using navigator respiratory gating methods.

Phase contrast techniques demonstrate directional blood flow information for improved identification of subtle intracardiac or extracardiac shunt lesions. Phase contrast also allows quantification of blood flow (e.g., estimation of the ratio of pulmonary to systemic blood flow [Qp/Qs]), regurgitant fractions, and pressure gradients across stenotic regions.

MRI has been used for diagnosing essentially all congenital heart and great-vessel abnormalities. It has been shown to have very high sensitivity and specificity for diagnosing common CHD. At a specificity of 90%, spin echo was found to have high sensitivity in diagnosing great-vessel relationships (100%), thoracic aortic abnormalities (94%), ASDs (91%), VSDs (100%), viscerotransposition (100%), and the cardiac loop (100%). Pulmonary and systemic venous anomalies and right ventricular outflow obstructions are also detected with high sensitivity. Vascular rings can also be accurately diagnosed without the need for angiography. MRI can also be performed using 3D techniques for high-spatial-resolution gadolinium-enhanced 3D MRA, or to provide volumetric coverage of cardiac chambers. Time-resolved MRA has been found to provide a very high diagnostic value (92% of diagnostic parameters assessed) that included thoracic vascular anatomy, sequential cardiac anatomy, and shunt detection with high sensitivity (93%-100%) and high specificity (87%-100%).

Gradient-echo imaging acquisition viewed in a cine format facilitates physiologic measurements, including stroke volume, ejection fraction, and wall motion of both ventricles. Blood flow, valve gradients, shunt flow, regurgitant flow, and pulmonary flow can all be measured using velocity-encoded cine techniques.

MRI seems to be ideally suited for evaluating adults with suspected or known CHD. Although claustrophobia in the gantry may require sedation in a few patients, the study is noninvasive, and image quality is not affected by body habitus. MRI can provide high-spatial-resolution images even in more complex CHD without the limitation of imaging "windows" or plane as experienced during echocardiography. MRI images can be obtained in essentially any plane for improved 3D presentation of cardiac anatomy. MRI is useful as well in evaluating the postoperative patients with CHD, whether it involves a palliative procedure, a surgically created conduit, or reconstructed great vessels.

MRI, however, does have a few contraindications and limitations. For instance, pacemakers are generally considered an exclusion for MRI, although it has been performed safely in patients with pacemakers under rigorously safe conditions. The use of gadolinium (Gd) chelate contrast agents may not be possible in a patient with known severe allergy to Gd. Detection of calcification remains problematic for MRI, so adults with homografts or bioprosthetic valved conduits in whom the detection of calcification implies deterioration may not be optimally imaged. Motion and respiratory artifacts also may pose a problem on some examinations.

In terms of specific defects, MRI is probably not as accurate as color flow Doppler in visualizing small ventricular and atrial defects. Detection of valvular pathology is perhaps better achieved with TEE. Cardiac MRI studies require supervision and monitoring of the procedure by a physician who understands the clinical question and can acquire an appropriate and optimal imaging study. This is essential for consistency and reliable data.

#### Transthoracic and Transesophageal Echocardiography versus Magnetic Resonance Imaging

Few prospective studies are available to compare TTE and TEE with MRI. Studies limited to specific congenital lesions (coarctation of the aorta, subpulmonary and pulmonary artery anomalies) indicate that MRI gives a more reliable assessment of severity and is technically more successful than TTE. Studies comparing TTE with MRI in the evaluation of patients who have had surgical correction or palliation of CHD indicate that MRI information is additive to that from TTE. In patients who have had palliative and corrective surgery for cyanotic heart disease, MRI and TEE are equivalent for demonstrating abnormalities of the right ventricular outflow tract, main pulmonary artery, and systemic-to-pulmonary shunts. MRI is superior in demonstrating abnormalities of the right and left pulmonary arteries. When TTE and MRI are compared for imaging a variety of congenital heart lesions, MRI is comparable to echocardiography in evaluating isolated intracardiac defects but more useful in diagnosing complex congenital lesions.

Echocardiography has also been shown to have good agreement with MRI in evaluating right ventricular volumes, but echocardiography has been noted to have a much wider interobserver variation. Similar agreement has been reported between echocardiographic and MRI measurements in patients with a functional single ventricle, with MRI showing better reproducibility than echocardiography in certain measurements such as myocardial mass and diastolic volume. In the evaluation of extracardiac ventriculopulmonary conduits and the right ventricle, MRI and

echocardiography can often provide complementary and diagnostic information that, when in agreement, may obviate the need for cardiac catheterization.

When TEE and MRI have been evaluated prospectively in adults with CHD, TEE is shown to be superior in evaluating intracardiac anatomy, but MRI is superior for extracardiac anatomy and is slightly better than TEE for hemodynamic and functional evaluation. Taken individually, the two modalities provided similar overall diagnostic information; however, when used in combination, they provide important complementary information in all diagnostic categories.

### Cardiac Catheterization and Angiocardiography

Cardiac catheterization has been the traditional gold standard for the diagnosis and management of CHD over the past 50 years. For the past 20 years, it has been increasingly supplemented by noninvasive diagnostic modalities—initially, cardiac ultrasound and more recently, CT scanning and MRI. Advances in these technologies have been logarithmic, and it is likely that in the coming decade, both morphologic and functional assessments of this patient population will be increasingly accomplished noninvasively.

In 2001, the 32<sup>nd</sup> Bethesda Conference Task Force 1, "The Changing Profile on Congenital Heart Disease in Adult Life," suggested the use of diagnostic catheterization primarily to resolve specific issues related to surgical intervention, (e.g., preoperative evaluation of coronary arteries, assessment of pulmonary vascular disease and its response to vasoactive agents for planned surgical intervention, and/or heart or heart/lung transplantation) and as an adjunct to noninvasive assessment of morphologic and/or functional characteristics of complex CHD (e.g., for delineating arterial and venous anatomy in patients with heterotaxy, patients who are candidates for a Fontan procedure, or patients who have had previous palliation in the form of a shunt). The group further suggested that only trained and experienced operators who maintain an adequate volume of such procedures annually should perform them. The Task Force further noted that evaluation for possible interventional catheterization is an increasingly common indication for diagnostic catheterization. Catheter intervention, for instance, is commonly sought as the treatment of choice for correcting valvular pulmonary stenosis, branch pulmonary stenosis, residual or recurrent aortic coarctation, and arteriovenous aortic coarctation, and arteriovenous fistulae. Coil or device occlusion of lesions such as patent ductus or secundum ASD are other preferred interventions for treatment.

For many years, the purpose of cardiac catheterization and angiocardiography for CHD was to acquire pressure, oximetric, and morphologic data. Pressures defined gradients across stenosis and between cardiac chambers connected by defects as well as the severity of pulmonary hypertension. Oxygen saturations helped to define the volume of shunts. Morphologic data of simple and complex anomalies were obtained by cine angiograms using angulated views, contrast material, and radiation. For the most part, these studies were accomplished safely but with some morbidity (contrast reactions, renal failure, hematomas, arterial and venous injuries, radiation exposure, etc.) and a small but definite mortality.

Although cardiac catheterization continues to be performed and is still considered by many to be the gold standard in evaluating CHD, noninvasive methods increasingly limit the need for catheterization unless intervention is considered. Many simple congenital cardiac defects are now sent to surgery without catheterization. In the future, cardiac catheterization and angiocardiography may very well be reserved as a complement to these noninvasive techniques in the evaluation of adults with suspected CHD. However, until these less invasive studies provide an accurate depiction of the coronary arteries, the catheterization laboratory will continue to be involved in the assessment of this unique group of adult patients.

### Summary

- TTE and MRI are frontline modalities for evaluating heart function and morphology and provide complementary information in diagnosing or assessing adult congenital heart disease.
- CT with contrast may be an alternative to MRI and TEE/TTE for evaluating heart function. The high spatial resolution of CT is helpful in evaluating smaller and tortuous vessels commonly seen in some disorders. CT is also advantageous in assessing the airway.
- Although noninvasive methods have reduced some of the need for cardiac catheterization, it still remains an important adjunct to noninvasive imaging for hemodynamic measurements and for interventions.
- MRA of the chest and CTA of the coronary arteries are important adjunctive modalities for evaluating associated vascular anomalies.

### Anticipated Exceptions

Nephrogenic systemic fibrosis (NSF) is a disorder with a scleroderma-like presentation and a spectrum of manifestations that can range from limited clinical sequelae to fatality. It appears to be related to both underlying severe renal dysfunction and the administration of gadolinium-based contrast agents. It has occurred primarily in patients on dialysis, rarely in patients with very limited glomerular filtration rate (GFR) (i.e., <30 mL/min/1.73 m<sup>2</sup>), and almost never in other patients. There is growing literature regarding NSF. Although some controversy and lack of clarity remain, there is a consensus that it is advisable to avoid all gadolinium-based contrast agents in dialysis-dependent patients unless the possible benefits clearly outweigh the risk, and to limit the type and amount in patients with estimated GFR rates <30 mL/min/1.73 m<sup>2</sup>. For more

information, please see the American College of Radiology (ACR) Manual on Contrast Media (see the "Availability of Companion Documents" field).

Abbreviations

- CT, computed tomography
- CTA, computed tomography angiography
- MRA, magnetic resonance angiography
- MRI, magnetic resonance imaging
- Tc-99m, technetium-99 metastable isomer
- TEE, transesophageal echocardiography
- TTE, transthoracic echocardiography
- US, ultrasound

Relative Radiation Level Designations

Relative Radiation Level*	Adult Effective Dose Estimate Range	Pediatric Effective Dose Estimate Range
O	0 mSv	0 mSv
<input type="text"/>	<0.1 mSv	<0.03 mSv
<input type="text"/> <input type="text"/>	0.1-1 mSv	0.03-0.3 mSv
<input type="text"/> <input type="text"/> <input type="text"/>	1-10 mSv	0.3-3 mSv
<input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/>	10-30 mSv	3-10 mSv
<input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/> <input type="text"/>	30-100 mSv	10-30 mSv
*RRL assignments for some of the examinations cannot be made, because the actual patient doses in these procedures vary as a function of a number of factors (e.g., region of the body exposed to ionizing radiation, the imaging guidance that is used). The RRLs for these examinations are designated as NS (not specified).		

Clinical Algorithm(s)

Algorithms were not developed from criteria guidelines.

Scope

Disease/Condition(s)

Congenital heart disease (CHD)

Guideline Category

Diagnosis

Evaluation

Clinical Specialty

Cardiology

Family Practice

Internal Medicine

Nuclear Medicine

Radiology

Thoracic Surgery

## Intended Users

Health Plans

Hospitals

Managed Care Organizations

Physicians

Utilization Management

## Guideline Objective(s)

To evaluate the appropriateness of initial radiologic examinations for adults with known or suspected congenital heart disease

## Target Population

Adults with known or suspected congenital heart disease

## Interventions and Practices Considered

1. X-ray, chest
2. Ultrasound (US) echocardiography
  - Transthoracic, resting
  - Transesophageal
3. Magnetic resonance imaging (MRI), heart function and morphology
  - Without and with contrast
  - Without contrast
4. Computed tomography angiography (CTA), coronary arteries
  - With contrast
  - With contrast with advanced low dose techniques
5. Computed tomography (CT), heart, with contrast
6. Magnetic resonance angiography (MRA), chest (noncoronary)
  - Without contrast
  - Without and with contrast
7. Cardiac catheterization with angiocardiology
8. Technetium (Tc)-99m ventriculography

## Major Outcomes Considered

Utility of radiologic examinations in differential diagnosis

## Methodology



## Methods Used to Collect/Select the Evidence

Searches of Electronic Databases

## Description of Methods Used to Collect/Select the Evidence

Literature Search Procedure

The Medline literature search is based on keywords provided by the topic author. The two general classes of keywords are those related to the condition (e.g., ankle pain, fever) and those that describe the diagnostic or therapeutic intervention of interest (e.g., mammography, MRI).

The search terms and parameters are manipulated to produce the most relevant, current evidence to address the American College of Radiology Appropriateness Criteria (ACR AC) topic being reviewed or developed. Combining the clinical conditions and diagnostic modalities or therapeutic procedures narrows the search to be relevant to the topic. Exploding the term "diagnostic imaging" captures relevant results for diagnostic topics.

The following criteria/limits are used in the searches.

1. Articles that have abstracts available and are concerned with humans.
2. Restrict the search to the year prior to the last topic update or in some cases the author of the topic may specify which year range to use in the search. For new topics, the year range is restricted to the last 5 years unless the topic author provides other instructions.
3. May restrict the search to Adults only or Pediatrics only.
4. Articles consisting of only summaries or case reports are often excluded from final results.

The search strategy may be revised to improve the output as needed.

## Number of Source Documents

The total number of source documents identified as the result of the literature search is not known.

## Methods Used to Assess the Quality and Strength of the Evidence

Weighting According to a Rating Scheme (Scheme Given)

## Rating Scheme for the Strength of the Evidence

Strength of Evidence Key

Category 1 - The conclusions of the study are valid and strongly supported by study design, analysis and results.

Category 2 - The conclusions of the study are likely valid, but study design does not permit certainty.

Category 3 - The conclusions of the study may be valid but the evidence supporting the conclusions is inconclusive or equivocal.

Category 4 - The conclusions of the study may not be valid because the evidence may not be reliable given the study design or analysis.

## Methods Used to Analyze the Evidence

Systematic Review with Evidence Tables

## Description of the Methods Used to Analyze the Evidence

The topic author drafts or revises the narrative text summarizing the evidence found in the literature. American College of Radiology (ACR) staff draft an evidence table based on the analysis of the selected literature. These tables rate the strength of the evidence for all articles included in the narrative text.

The expert panel reviews the narrative text, evidence table, and the supporting literature for each of the topic-variant combinations and assigns an appropriateness rating for each procedure listed in the table. Each individual panel member forms his/her own opinion based on his/her interpretation of the available evidence.

More information about the evidence table development process can be found in the American College of Radiology (ACR) Appropriateness Criteria® Evidence Table Development document (see the "Availability of Companion Documents" field).

## Methods Used to Formulate the Recommendations

Expert Consensus (Delphi)

## Description of Methods Used to Formulate the Recommendations

Modified Delphi Technique

The appropriateness ratings for each of the procedures included in the Appropriateness Criteria topics are determined using a modified Delphi methodology. A series of surveys are conducted to elicit each panelist's expert interpretation of the evidence, based on the available data, regarding the appropriateness of an imaging or therapeutic procedure for a specific clinical scenario. American College of Radiology (ACR) staff distributes surveys to the panelists along with the evidence table and narrative. Each panelist interprets the available evidence and rates each procedure. The surveys are completed by panelists without consulting other panelists. The ratings are a scale between 1 and 9, which is further divided into three categories: 1, 2, or 3 is defined as "usually not appropriate"; 4, 5, or 6 is defined as "may be appropriate"; and 7, 8, or 9 is defined as "usually appropriate." Each panel member assigns one rating for each procedure per survey round. The surveys are collected and the results are tabulated, de-identified and redistributed after each round. A maximum of three rounds are conducted. The modified Delphi technique enables each panelist to express individual interpretations of the evidence and his or her expert opinion without excessive bias from fellow panelists in a simple, standardized and economical process.

Consensus among the panel members must be achieved to determine the final rating for each procedure. Consensus is defined as eighty percent (80%) agreement within a rating category. The final rating is determined by the median of all the ratings once consensus has been reached. Up to three rating rounds are conducted to achieve consensus.

If consensus is not reached, the panel is convened by conference call. The strengths and weaknesses of each imaging procedure that has not reached consensus are discussed and a final rating is proposed. If the panelists on the call agree, the rating is accepted as the panel's consensus. The document is circulated to all the panelists to make the final determination. If consensus cannot be reached on the call or when the document is circulated, "No consensus" appears in the rating column and the reasons for this decision are added to the comment sections.

## Rating Scheme for the Strength of the Recommendations

Not applicable

## Cost Analysis

A formal cost analysis was not performed and published cost analyses were not reviewed.

## Method of Guideline Validation

Internal Peer Review

## Description of Method of Guideline Validation

Criteria developed by the Expert Panels are reviewed by the American College of Radiology (ACR) Committee on Appropriateness Criteria.

# Evidence Supporting the Recommendations

## Type of Evidence Supporting the Recommendations

The recommendations are based on analysis of the current literature and expert panel consensus.

## Benefits/Harms of Implementing the Guideline Recommendations

### Potential Benefits

Selection of appropriate radiologic imaging procedures for known or suspected congenital heart disease

### Potential Harms

- There is a small risk of bacterial endocarditis from transesophageal echocardiography (TEE).
- Detection of calcification is a problem for magnetic resonance imaging (MRI), so adults with homografts or bioprosthetic valved conduits in whom the detection of calcification implies deterioration may not be optimally imaged. Motion and respiratory artifacts also may pose a problem on some examinations.

### Relative Radiation Level (RRL)

Potential adverse health effects associated with radiation exposure are an important factor to consider when selecting the appropriate imaging procedure. Because there is a wide range of radiation exposures associated with different diagnostic procedures, a relative radiation level indication has been included for each imaging examination. The RRLs are based on effective dose, which is a radiation dose quantity that is used to estimate population total radiation risk associated with an imaging procedure. Patients in the pediatric age group are at inherently higher risk from exposure, both because of organ sensitivity and longer life expectancy (relevant to the long latency that appears to accompany radiation exposure). For these reasons, the RRL dose estimate ranges for pediatric examinations are lower as compared to those specified for adults. Additional information regarding radiation dose assessment for imaging examinations can be found in the American College of Radiology (ACR) Appropriateness Criteria® Radiation Dose Assessment Introduction document (see the "Availability of Companion Documents" field).

### Gadolinium-based Contrast Agents

Nephrogenic systemic fibrosis (NSF) is a disorder with a scleroderma-like presentation and a spectrum of manifestations that can range from limited clinical sequelae to fatality. It appears to be related to both underlying severe renal dysfunction and the administration of gadolinium-based contrast agents. It has occurred primarily in patients on dialysis, rarely in patients with very limited glomerular filtration rate (GFR) (i.e.,  $<30$  mL/min/1.73 m<sup>2</sup>), and almost never in other patients. Although some controversy and lack of clarity remain, there is a consensus that it is advisable to avoid all gadolinium-based contrast agents in dialysis-dependent patients unless the possible benefits clearly outweigh the risk, and to limit the type and amount in patients with estimated GFR rates  $<30$  mL/min/1.73 m<sup>2</sup>. For more information, please see the American College of Radiology (ACR) Manual on Contrast Media (see the "Availability of Companion Documents" field).

## Contraindications

### Contraindications

- Pacemakers are generally considered an exclusion for magnetic resonance imaging (MRI), although it has been performed safely in patients with pacemakers under rigorously safe conditions.
- The use of gadolinium (Gd) chelate contrast agents may not be possible in a patient with known severe allergy to Gd.

## Qualifying Statements

## Qualifying Statements

The American College of Radiology (ACR) Committee on Appropriateness Criteria and its expert panels have developed criteria for determining appropriate imaging examinations for diagnosis and treatment of specified medical condition(s). These criteria are intended to guide radiologists, radiation oncologists, and referring physicians in making decisions regarding radiologic imaging and treatment. Generally, the complexity and severity of a patient's clinical condition should dictate the selection of appropriate imaging procedures or treatments. Only those examinations generally used for evaluation of the patient's condition are ranked. Other imaging studies necessary to evaluate other co-existent diseases or other medical consequences of this condition are not considered in this document. The availability of equipment or personnel may influence the selection of appropriate imaging procedures or treatments. Imaging techniques classified as investigational by the U.S. Food and Drug Administration (FDA) have not been considered in developing these criteria; however, study of new equipment and applications should be encouraged. The ultimate decision regarding the appropriateness of any specific radiologic examination or treatment must be made by the referring physician and radiologist in light of all the circumstances presented in an individual examination.

## Implementation of the Guideline

### Description of Implementation Strategy

An implementation strategy was not provided.

## Institute of Medicine (IOM) National Healthcare Quality Report Categories

### IOM Care Need

Living with Illness

### IOM Domain

Effectiveness

## Identifying Information and Availability

### Bibliographic Source(s)

Ho VB, Biko DM, White RD, Woodard PK, Abbara S, Dorbala S, Haramati LB, Mammen L, Martin ET III, Rigsby CK, Ryan T, White CS, Expert Panel on Cardiovascular Imaging. ACR Appropriateness Criteria® known or suspected congenital heart disease in the adult. [online publication]. Reston (VA): American College of Radiology (ACR); 2011. 9 p. [80 references]

### Adaptation

Not applicable: The guideline was not adapted from another source.

### Date Released

1998 (revised 2011)

## Guideline Developer(s)

American College of Radiology - Medical Specialty Society

## Source(s) of Funding

The American College of Radiology (ACR) provided the funding and the resources for these ACR Appropriateness Criteria®.

## Guideline Committee

Committee on Appropriateness Criteria, Expert Panel on Cardiovascular Imaging

## Composition of Group That Authored the Guideline

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## Financial Disclosures/Conflicts of Interest

Not stated

## Guideline Status

This is the current release of the guideline.

This guideline updates a previous version: Ho VB, Yucel EK, Khan A, Haramati LB, Mammen L, Rozenshtein A, Rybicki FJ, Schoepf UJ, Stanford W, Stein B, Woodard PK, Jaff M, Expert Panel on Cardiovascular Imaging. Suspected congenital heart disease in the adult. [online publication]. Reston (VA): American College of Radiology (ACR); 2007. 8 p.

The appropriateness criteria are reviewed biennially and updated by the panels as needed, depending on introduction of new and highly significant scientific evidence.

## Guideline Availability

Electronic copies: Available from the [American College of Radiology \(ACR\) Web site](#) .

Print copies: Available from the American College of Radiology, 1891 Preston White Drive, Reston, VA 20191. Telephone: (703) 648-8900.

## Availability of Companion Documents

The following are available:

- ACR Appropriateness Criteria®. Overview. Reston (VA): American College of Radiology; 2 p. Electronic copies: Available in Portable Document Format (PDF) from the [American College of Radiology \(ACR\) Web site](#) .
- ACR Appropriateness Criteria®. Literature search process. Reston (VA): American College of Radiology; 1 p. Electronic copies: Available in Portable Document Format (PDF) from the [ACR Web site](#) .
- ACR Appropriateness Criteria®. Evidence table development – diagnostic studies. Reston (VA): American College of Radiology; 2013 Nov. 3 p. Electronic copies: Available in PDF from the [ACR Web site](#) .
- ACR Appropriateness Criteria®. Radiation dose assessment introduction. Reston (VA): American College of Radiology; 2 p. Electronic copies: Available in Portable Document Format (PDF) from the [ACR Web site](#) .

- ACR Appropriateness Criteria® Manual on contrast media. Reston (VA): American College of Radiology; 90 p. Electronic copies: Available in PDF from the [ACR Web site](#) .
- ACR Appropriateness Criteria® known or suspected congenital heart disease in the adult. Evidence table. Reston (VA): American College of Radiology; 2011. 22 p. Electronic copies: Available from the [ACR Web site](#) .

## Patient Resources

None available

## NGC Status

This summary was completed by ECRI on February 20, 2001. The information was verified by the guideline developer on March 14, 2001. This summary was updated by ECRI on March 31, 2003. The updated information was verified by the guideline developer on April 21, 2003. This NGC summary was updated by ECRI Institute on November 12, 2007. This summary was updated by ECRI Institute on January 13, 2011 following the U.S. Food and Drug Administration (FDA) advisory on gadolinium-based contrast agents. This summary was updated by ECRI Institute on July 8, 2011.

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